Recent Advances in Surgery of Congenital Heart Disease

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■ *In the cyanotic group palliative procedures for transposition of the great arteries are frequently life-saving in infancy, and the definitive operations such as the atrial baffle, and the Rastelli procedure for those with ventricular septal defect and pulmonic stenosis, are now firmly established. In tetralogy of Fallot shunting procedures continue to be employed in infancy and early childhood, and the complete repair is usually done after the age of five. Corrective operations for total anomalous venous return may have to be staged, and the results are more satisfactory in older children. The various forms of endocardial cushion defects can usually be recognized accurately preoperatively, and where the normal anatomical relationships can be restored, excellent results obtained. Brilliant operative success can now be had in some forms of truncus arteriosus and double outlet right ventricle.*

It is quite common to find congenital heart disease in adults, frequently after many years of having been treated as rheumatic heart disease. The operative risk in this group is less than 10 percent, and in most instances such patients are restored to their normal physiological age after operation.

REMARKABLE ADVANCES have been made in the surgical treatment of congenital cardiac malformations in the past decade. Many of the old procedures have established a permanent place in surgical therapy, while new operations are being devised for hitherto uncorrectable complex lesions. If the 1940s are to be remembered for the beginning of surgical correction of extracardiac lesions (patent ductus arteriosus and coarctation) and the 1950s for the introduction of extracorporeal circulation, the past decade will be remembered for the improvement in mortality statistics as surgeons continue to learn more about preoperative, intraoperative, and postoperative care of the patient with congenital heart disease.

Healthy development in this field is noted in attempts to establish classification, nomenclature, criteria for diagnosis (New York Heart Association, 1964)[1] development of a method of coding

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TABLE 1.—*Procedures Used in Treatment of Transposition of the Great Arteries*

Palliative:
1. Creation of atrial septal defect (Blalock and Hanlon, 1949).
2. Baffes' operation (1956).
3. Balloon septostomy (Rashkind et al, 1966).
4. Edwards' procedure (1966).

Corrective:
1. Correction at the atrial level (Senning, 1959 and Mustard, 1964).
2. Correction at the arterial level (Kay, 1955).
3. Correction at the ventricular level (Rastelli, et al, 1969).

TABLE 2.—*Results of Mustard Operation*[11]

	Type I Transposition with Intact Septum		*Type II with VSD Transposition*		*Type III Transposition, VSD and Pulmonary Stenosis*	
Author	*Patients*	*Mortality (Percent)*	*Patients*	*Mortality (Percent)*	*Patients*	*Mortality (Percent)*
Aberdeen	35	14	13	15	1	100
Cooley	9	28	5	80	8	75
Kirklin	6	16	13	30	2	50
Mustard	18	29	6	84	4	100

for data processing systems (Kerth et al)[2] and cooperative studies involving various centers (Kittle, 1968)[3] regarding "categorizing and defining quantitatively the preoperative observations which are predictive of risk and quality of survival."

The purpose of this communication is to review briefly the new developments in the surgical therapy of congenital heart disease and also to analyze the results of operation for common congenital cardiac defects from various centers including our own. The shortness of this review necessitates exclusion of many excellent series. Adequate description of the procedures or acknowledgement of the contributions from many distinguished contributors is impossible for the same reason.

Cyanotic Group

Transposition of the Great Arteries

Transposition of the great arteries is the commonest cause of mortality in infants born with congenital heart disease. Eighty-six percent of these children die during the first six months of life. As the operations for this anomaly have to be performed in very small infants, the surgical risk is extremely high. In recent years, a number of palliative and corrective procedures have been available since the creation of an atrial septal defect was suggested by Blalock and Hanlon.[4] (See Table 1.)

Among the palliative procedures, the Blalock-Hanlon operation has been the most commonly employed, even though the lowest mortality figure reported with this operation is 18 percent (Ochsner et al).[5] In the future, however, balloon septostomy, as described by Rashkind,[6] will be used more frequently because of the low mortality associated with it.

Until recently, the reports of successful total correction of transposition of the great arteries were few and isolated (Senning,[7] Kirklin[8]). In 1964, Mustard[9] described a method of correction at the atrial level, using a pericardial baffle on a principle described originally by Albert.[10] Subsequent use of this procedure by other surgeons was very prompt and their results are compared in Table 2.[11] For transposition with pulmonic stenosis and ventricular septal defect, the results of the Mustard operation are not so satisfactory. Rastelli et al, 1969[12] recently devised a new method of correction at the ventricular level and used it successfully in several children. The reader is referred to their excellent paper for the details of the correction, which basically consists in using a prosthetic baffle between the ventricular septal defect and the aortic orifice so as to divert the flow of blood from the left ventricle to aorta. An aortic homograft or autologous fascia lata graft is used to reconstruct a new right ventricular outflow tract from the anatomic right chamber.

Tetralogy of Fallot

Surgical treatment for tetralogy of Fallot is 25 years old, a quarter century having passed since Blalock and Taussig devised a method of systemic-pulmonary anastomosis for this anomaly which comprises 30 percent of the cyanotic group and 11 percent of all congenital cardiac malformations. Table 3 lists a number of palliative and corrective procedures in use for tetralogy. The variation in the degree of outflow obstruction of the right ventricle in tetralogy accounts for such a variety of surgical procedures and the need for surgery at the different levels.

Early palliative shunting procedures are required for many of the severely cyanotic infants. In the infants who have anoxic difficulties before the age of two months, the outlook may be poor because of atretic pulmonary vessels; however, it is frequently possible to perform a systemic-pulmonary artery anastomosis even in small infants. It is usually technically more satisfactory to use

TABLE 3.—*Surgical Procedures in Use for Tetralogy of Fallot*

Palliative:	Corrective:
Systemic-Pulmonary shunts	• Lillehei (1955)
• Blalock-Taussig (1945)	
• Potts (1946)	
• Glenn (1956)	
• Waterston (1962)	
Direct attack on pulmonic valve	
• Brock (1948)	

TABLE 4.—*Surgical Mortality of Blalock-Taussig Operation*

Author	*Number of Cases*	*Mortality (Percent)*
Taussig-Bauersfeld (1953)	857	15.0
Möller (1962)	148	14.9
Hallman-Cooley (1963)	205	8.3
Gerbode (1963)	132	8.5
Shumacker-Mandelbaum (1960)	115	4.3
Sulamma (1964)	51	3.9

the subclavian artery arising from the innominate.[13] The results of this operation are summarized in Table 4. The Potts[14] type of anastomosis and the Glenn[15] procedure are not as frequently used because of the technical difficulty at the time of total correction. Some surgeons prefer the Waterston type of shunt, between the ascending aorta and right pulmonary artery.[16]

Most surgeons prefer not to operate for total correction in children less than five years of age, although others are ready to lower this age limit to three. The surgical mortality for total correction has improved considerably since Lillehei's[17] first attempt under cross circulation. This certainly can be attributed to better understanding of the anatomic features of the lesion and the conduction system of the heart, and to greatly improved methods of perfusion and postoperative care. (See Table 5.)

Anomalous Pulmonary Venous Drainage

This anomaly, which frequently is fatal during the first year of life, consists of all the pulmonary veins opening into the right atrium by means of a left superior vena cava or various other partial or total venous return to the right atrium, coronary sinus, or inferior vena cava. In total anomalous venous return, the systemic distribution of blood occurs through a patent foramen ovale. Often in this condition congestive failure develops in early infancy. The corrective operation for this abnormality consists of the anastomosis of the left

TABLE 5.—*Surgical Mortality of Total Correction of Tetralogy of Fallot*

Author	*Number of Cases*	*Mortality (Percent)*
Kirklin (1965)	509	7.0
Kimoto (1965)	72	19.0
Zenker (1964)	216	24.0
Zerbini (1965)	221	13.5
Gerbode (1963)	75	13.0
Kay (1959)	50	18.0
Barnard & Schrire (1961)	42	17.0
Mustard (1962)	188	13.0
Shumway (1965)	44	
Malm (1963)	41	

atrium to the common pulmonary vein which lies behind it, and ligation of the left superior vena cava. As some of the patients have a hypoplastic left heart, Mustard[18] suggested delaying closure of the atrial septal defect in these cases. Another method we have used is delayed ligation of the left superior vena cava. Pulmonary edema is the leading cause of death in such patients after surgical repair. The results of operation for partial anomalous venous return are excellent. The mortality rate for surgical operation in total anomalous return in infants is 53 percent,[19] whereas in the older age group the prognosis is much better. Occasionally, one finds adults who have escaped diagnosis of total anomalous venous return during childhood, but who can be operated upon successfully. One of these had served in the infantry in the last European war.

Rare Anomalies

Among the less common cyanotic conditions, new methods of corrections have been suggested for tricuspid and pulmonary atresia, and Ebstein's malformation of tricuspid valve. The superior vena cava to right pulmonary artery anastomosis[15] has provided good temporary palliation in these cases.

Tricuspid atresia. Patients with tricuspid atresia are cyanotic, have left ventricular hypertrophy, and a heart of relatively normal size because of the atretic tricuspid valve and hypoplastic right ventricle. Operation is usually required early in infancy because of the high mortality during the first year of life in untreated cases. Excellent palliation can be obtained by systemic-pulmonary artery shunt or the Glenn operation, but recently Rams et al (1966)[20] suggested an operation which seems quite imaginative. They described a three-stage correction for this condition: Glenn operation in the first stage, an anastomosis between right

atrial appendage and main pulmonary artery as the second stage and closure of the atrial septal defect in the third and final stage.

Pulmonary atresia. Pulmonary atresia with normal aortic root is another uncommon but challenging anomaly because of the mortality rate of 80 percent during the first year of life. In this condition the pulmonary valve and artery are hypoplastic and the right ventricle rudimentary. Life is maintained by a patent ductus arteriosus, closure of which accounts for the shortened life span. Shunting procedures are feasible but mortality is high. Campbell[21] collected reports of 27 cases and noted good results in only 25 percent. Four patients in his own purview went on to survive to the ages of three, five, eight, and ten years with shunt operations. Sometimes it is better to do both caval and systemic shunts in these patients.

Ebstein's anomaly. In this malformation there is abnormal displacement of tricuspid valve into the right ventricle. In correcting it, one has to take into consideration the conditions of valve leaflets and atrialized right ventricle distal to the valve. If the valve is fairly normal, it is possible to return the valve and its annulus to the annulus fibrosis with the excision of atrialized portion of right ventricle, as done by Hardy and his associates.[22] However, if the valve leaflets are severely deformed they have to be removed and a prosthetic valve inserted.[23]

Acyanotic Group

Septal Defects

Ventricular septal defect. Ventricular septal defects with pulmonary hypertension constitute a major challenge during infancy, whereas mortality for operating upon defects with normal pulmonary artery pressure is approaching zero. Most infants with ventricular septal defects can be managed by medical regimen during the first year of life, while some may require pulmonary artery banding because of high pulmonary flow. This procedure carries an acceptably low mortality and allows the surgeon a period of two to three years for definitive open heart repair.[24] In the presence of severe left ventricular failure, progressive pulmonary vascular disease and severe growth failure, an early intracardiac repair of the ventricular septal defect may be preferable to pulmonary artery banding. An ideal candidate for intracardiac repair, however, is a child more than five years old with large left-to-right shunt with a slight to moderate elevation of right ventricular pressure. We believe that virtually all ventricular septal defects with shunts above 2 to 1 should be closed by the age of ten, since the chances of spontaneous closure beyond that age are minimal.

TABLE 6.—*Surgical Mortality: Endocardial Cushion Defects—Partial A-V Canal*

Author	*Number of Cases*	*Mortality (Percent)*
Scott (1962)	32	19
Mustard (1965)	61	16
McGoon (1959)	35	6
Barnard (1968)	28	7
Gerbode (1967)	39	5

TABLE 7.—*Surgical Mortality: Endocardial Cushion Defects—Complete A-V Canal*

Author	*Number of Cases*	*Mortality (Percent)*
Mustard (1965)	23	73
McGoon (1959)	15	73
Scott (1962)	12	67
Barnard (1968)	6	33
Gerbode (1967)	29	30

Atrial septal defects. Closure of an atrial septal defect of secundum type is one of the most gratifying operations in cardiac surgery. The mortality is less than 2 percent.[25,26] We recommend operation in all children five years of age or older who have shunts greater than 1.5 to 1.

Endocardial cushion defects. This term was introduced by Watkins and Gross.[27] In includes a group of defects previously called partial or complete atrioventricular canal. Our preference is, however, for the classification of Paul[28]: ostium primum defect, with cleft mitral valve, with cleft tricuspid and mitral valves, and atrioventricular communis with ventricular septal defect and mitral and tricuspid valves appearing as common valve. Our preferred method of repair of the different types is described elsewhere.[29] The mortality associated with operating upon patients with complete A-V canal is understandably high because of the anatomical complexities, and low in the partial A-V canal group. The high mortality in the former group (Tables 6 and 7) is mainly due to the lack of satisfactory cusp tissue (as a result of which neither the valves nor the defect can be repaired), surgical heart block, and frequent association of other anomalies. Patients who survive operation are generally greatly improved or, in fact, completely cured.

Stenotic Valve Lesions

Valvular pulmonic stenosis with intact ventricular septum represents about 10 percent of congenital abnormalities. In infancy, it constitutes a major threat to life if not recognized and treated early. Infants with one anoxic episode or in congestive failure require immediate operation, while older children with gradients across the pulmonary valve can be operated upon electively. The operative approach is either transventricular or transarterial via pulmonary artery. Although blind valvotomy and the open operation utilizing inflow occlusion and hypothermia have been employed, it is our preference to perform the open procedure with the aid of extracorporeal circulation in nearly all instances, as the operative risk is less and the result more satisfactory.

Congenital Aortic Stenosis

This condition may be valvular, subvalvular, supravalvular or a combination of these. Severe valvular stenosis can be the cause of sudden death in childhood, but the usual symptoms are fatiguability, exertional dyspnea, angina pectoris and syncope. Though the valvular stenosis is the most common, muscular subaortic stenosis is the lesion which has been the subject of recent clinical curiosity and investigation.[30] The results of surgical treatment in muscular subaortic stenosis are very satisfactory. Although several methods of relieving the obstruction have been suggested we have found that very satisfactory results, with no mortality, have been obtained with a method similar to that described by Trimble[31] and Morrow.[32] For valvular stenosis, careful commissurotomy is necessary and residual gradients are common. For supravalvular stenosis a prosthetic gusset is used for widening the orifice. At present there is no acceptable operation for hypoplastic aortic annulus.

Miscellaneous Congenital Anomalies

Persistent Truncus Arteriosus

In recent years there have been brilliant reports of successful operations for correction of complex anomalies which were previously considered untreatable. Persistent truncus arteriosus is a condition in which a pulmonary artery arises from a single aortic vessel, the truncus, where it leaves the base of the heart. Also there is a high ventricular septal defect. This single trunk supplies the coronary and the systemic and pulmonary circulation.

TABLE 8.—*Results of Operation in Infants with Congenital Heart Disease Under the Age of Two Years*

Author	*Number of Cases*	*Mortality (Percent)*
Zerbini (1964)	71	41
Thorkelsen (1964)	200	33
Cooley (1964)*	500	27
Gerbode (1964)	147	25
Aberdeen (1968)*	835	32

*Under one year of age.

TABLE 9.—*Mortality During the First Year of Life—(From Various Autopsy Series)*

Congenital Cardiac Lesion	*Percentage Dying Under One Year of Age—(Percent)*
Pulmonary atresia	100
Transposition of great vessels	85
Tricuspid atresia	83
Total anomalous pulmonary venous drainage	80
Pulmonary stenosis	70
Coarctation of aorta	75
Tetralogy of Fallot	45
Ventricular septal defect	42

TABLE 10.—*Results of Operation in Adults Over 21 with Congenital Cardiac Lesions*

	No. patients	*Hospital Mortality*
Open Heart Operations		
Atrial septal defect (secundum)	126	6 (4.7%)
Endocardial cushion defects	16	2 (12.4%)
Tetralogy of Fallot	28	6 (21%)
Ventricular septal defects	21	1 (5%)
Pulmonary Stenosis	18	0
Ruptured aneurysms of the sinus of Valsalva	9	1 (11%)
Left ventricular outflow tract obstruction other than valvular aortic stenosis	12	2 (16.6%)
Miscellaneous	10	4
Subtotal	240	22 (9.1%)
Closed Procedures		
Patent ductus arteriosus	29	0
Coarctation of the aorta	33	2 (6%)
Congenital heart block	1	0
Subtotal	63	2 (3.2%)
Total	303	24 (8%)

Fifty percent of the children born with this anomaly are dead within the first six months of life and survival up to early adult life is possible only if the pulmonary arteries are small. Pulmonary artery banding is used as temporary palliation but the results are not satisfactory. Recently Wallace et al (in 1968)[33] and Weldon (in 1968)[34] devised a method of successful total correction. This consists of the closure of the ventricular septal defect and the reconstruction of a new right ventricular outflow tract and main pulmonary artery, utilizing a homograft ascending aorta with its aortic valve.

TABLE 11.—*Surgical Classification of Congenital Heart Disease According to Operability*

I. Operable or Surgically Correctable Lesions
- A. Acyanotic Group
  - 1. Lesions With Abnormal Pulmonary-Systemic Shunts
    - (a) Excellent results as surgery is low risk
      - Patent ductus arteriosus
      - Atrial septal defect
      - Ventricular septal defect
      - Aorto-pulmonary window
      - Pulmonic stenosis with ASD
      - Pulmonic stenosis with VSD (acynotic tetrology)
      - Coronary arteriovenous fistula
      - Ruptured aortic sinuses of Valsalva
      - Partial atrioventricular canal
      - Ebstein's anomaly with ASD and left to right shunt
      - Left ventricle-right atrial communications
      - Aberrant pulmonary artery
    - (b) High risk associated with operation
      - Complete atrioventricular canal
      - Large ventricular septal defect with pulmonary hypertension during infancy
      - Preductal coarctation of aorta
  - 2. Lesions Without Abnormal Pulmonary-Systemic Shunts
    - (a) Excellent results as operation is low risk
      - Coarctation of aorta
      - Pulmonic stenosis (isolated)
      - Congenital aortic stenosis
      - Anomalies of the aortic arch
      - Anomalies of the coronary arteries
    - (b) High risk associated with operation
      - Tricuspid stenosis and atresia without ASD
      - Ebstein's malformation without ASD
      - Congenital mitral stenosis
      - Cor triatriatum
      - Ectopia cordis
      - Congenital diverticulum of the left ventricle
- B. Cyanotic Group
  - 1. Lesions Resulting in Decreased Pulmonary Blood Flow
    - (a) Excellent results as operation is low risk
      - Pulmonic stenosis as with ASD with right-to-left shunt
      - Tetralogy of Fallot
      - Pentalogy of Fallot
      - Abnormalies of cava: IVC or SVC to left atrium
    - (b) High risk associated with operation
      - Pulmonary atresia
      - Tricuspid atresia
      - Persistent truncus arteriosus with hypoplastic pulmonary arteries
      - Ebstein's anomaly with ASD and right-to-left shunt
  - 2. Lesions Resulting in Increased Pulmonary Blood Flow
    - (a) Excellent results as operation is low risk
      - Partial anomalous pulmonary venous return
      - Total anomalous pulmonary venous return with normal pulmonary vascular resistance
      - Transposition of great vessels with intact ventricular septum
      - Congenital pulmonary arteriovenous fistula
    - (b) High risk associated with operation
      - Truncus arteriosus
      - Double outlet right ventricle
      - Transposition of great arteries with VSD or pulmonic stenosis

II. Nonoperable or Surgically Uncorrectable Lesions
- Single ventricle
- Hypoplastic left heart syndrome
  - Mitral atresia
  - Aortic atresia
  - Atresia of aortic arch
  - Hypoplasia of the aorta
- Complete atrioventricular canal with severe deficiency of valve tissue
- Tricuspid and pulmonary atresia with transposition of great arteries
- Truncus arteriosus with severely hypoplastic pulmonary arteries
- Taussig-Bing anomaly

III. Lesions In Which Operation Is Contraindicated
- Endocardial fibroelastosis
- Primary pulmonary hypertension
- Eisenmenger's syndrome: any lesion or combination of lesions in which shunt is reversed due to pulmonary circulatory obstruction
- Von Gierke's disease
- Idiopathic hypertrophy of heart (Familial cardiomyopathy)

IV. Lesions In Which Operation Is Usually Not Necessary
- Dextrocardia
- Dextro-rotations
- Corrected transpositions

Double Outlet Right Ventricle

Double outlet right ventricle, a rare but interesting anomaly, was formerly considered inoperable. In essence it is a variety of incomplete transposition in which the anterior-posterior relation of the great arteries may be normal but the aorta originates from the right ventricle. The only outlet to the left ventricle is through the ventricular septal defect. The correction described by Redo et al in 1963[35] and Kirklin in 1964[36] consists in the use of a prosthetic or tissue baffle so as to provide a tunnel between the ventricular septal defect and the aortic orifice. Usually the ventricular septal defect needs enlarging. With operation the outlook is quite encouraging, for the life expectancy is poor in infancy. (In all 13 cases in the autopsy series from Johns Hopkins the patients were under six months.)

Cardiac Surgery in Infants

Although the surgical mortality in the first two years of life was very high in early days of cardiac surgery there has been steady improvement due not only to advancements in technique and postoperative care, but equally to the current accuracy in diagnosis. (See Table 8.) Today it is customary to perform cardiac catheterization and angiography at any hour on an emergency basis. It must be

emphasized also that the first year of life is extremely critical for these infants. Table 9 gives the percentage of cause of death of infants lost under the age of 12 months.

Congenital Heart Disease in Adults

Until fairly recently surgical treatment for the lesions of congenital heart disease often was delayed in adults in the belief that the patient had rheumatic heart disease. Greater use of laboratory diagnostic aids and more widespread recognition of clinical signs has changed this. Table 10 shows our operative experience in patients with congenital heart disease over the age of 21 years.[37] From the surgical results, it would be fair to conclude that in the absence of categorical contraindications, an adult with congenital heart disease is a proper candidate for surgical correction. Most of those who have complete repair are restored to approximately their normal physiological age.

On reviewing the experience in the past 30 years, it is possible to propose a classification on a surgical basis. The only possible use for such a classification is to outline what has been done and what remains to be done for the surgeon in this field. (See Table 11.)

In conclusion, the results of surgical repair for congenital heart disease have improved significantly in the past decade and many new procedures have become established for the correction of complex congenital abnormalities of heart for which only palliative operation was formerly available. In general, outlook for the future in this field is quite hopeful and one can safely predict further improvement in the results of operation because of our better understanding of the physiologic and anatomic features involved, as well as improvements in extracorporeal circulation and postoperative care.

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